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SUPPURATING CONGENITAL CYSTIC KIDNEY, WITH HYDRONEPHROSIS AND OBLITERATION OF THE URETER.

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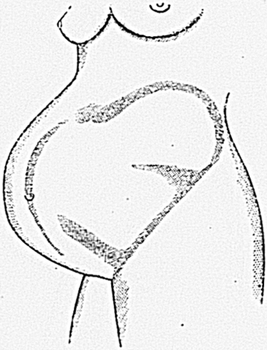
A CAREFUL search through medical literature would seem to warrant the assertion that the following case is unique, for although several cases have been reported in which hydronephrosis has been found associated with other pathological conditions of the kidney, no parallel to the instance in question could be discovered. Aside from its pathological importance, the case is of especial interest on account of the enormous size of the tumor, extending from roof to floor of the abdominal cavity, as well as its close resemblance to an ovarian cystoma, for which it was at first mistaken.

Finally, the successful operation for the removal of such a monstrous kidney, followed by the rapid convalesence of the patient, to-day enjoying the best of health, complete the interesting picture.

HISTORY.—*Outline.* Mrs. R. W., a spare little woman, of a markedly cachectic appearance, aged fifty-three years, a native of Seaford, Delaware. Admitted to Dr. H. A. Kelly's service in the Johns Hopkins Hospital, March 25, 1890. Health had been good until seven years ago, when she began to suffer with numerous attacks of fever and chills, diagnosticated as malarial. An abdominal tumor was discovered six years ago, and since then it had gradually increased to its present size.

FIG. 1.



Taken from a photograph. Shows the extreme prominence of the abdomen caused by the renal tumor, closely simulating large ovarian tumor.

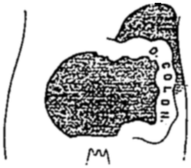
Detailed history. Her father died of "heat stroke," and her mother of Bright's disease, one uncle also died from Bright's disease, and one sister died in "coma." At the present time she has a cousin who suffers from a doubtful renal tumor.

She passed the menopause at the age of forty-nine, the menses having previously been regular and normal. She is V-para, eldest twenty-nine, youngest eleven years. One miscarriage. She has never had any trouble with the bowels, any urinary difficulty, or any previous serious illness. During her pregnancies she was always troubled with excessive nausea.

Her family physician states that "she had frequent attacks of intermittent chills and fever seven years ago, which were controlled by quinine and mercurials, and about the same time she began to have attacks of pain in the left side, which increased in frequency and severity up to date of operation. During these attacks the most comfortable position was "on the hard floor, with room to roll." Up to the time of admission to the hospital she had acquired the habit of taking large quantities of opium to relieve these pains. About six months previously she had congestive rigors.

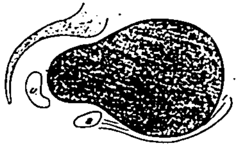
Six years ago she noticed a lump in the right groin, above Poupart's ligament. This was painful at times. It grew gradually until two years ago, when it increased rapidly to its present size within five months; since then it has remained stationary. The following are the notes of the examination on admission to the Hospital:

FIG. 2.



Shows the tumor in black, lying behind the colon and extending from the left hypochondrium to the floor of the pelvis.

FIG. 3.



Shows the tumor in sagittal section. Its relations to the anterior abdominal walls, the promontory, and the small depressed uterus are shown.

Description and mensuration of tumor. The abdomen prominent, vaulted, filled by an irregular bossed mass, about the size of a pregnant uterus at term. Two-thirds of this mass lies to the left of median line. The greatest girth is at umbilicus 104 cm. (40 in.). From ensiform cartilage to symphysis pubis 33 cm. (14 in.). Right spine of ilium to umbilicus 21 cm. (8 in.). Left spine of ilium to umbilicus 26 cm. (10 in.). On the right is a hemispherial fluctuant mass like a monocyst, which is 21 cm. (8 in.) in vertical, and 10 cm. (3¾ in.) in transverse measurement. The intestines are plainly felt, and percussed in right flank. To the left the tumor is made up of three large bosses, one just over median line corresponding to the one on right, one above this, and one filling out the left flank. Small intestines pushed far back, but on percussion there is *a remarkable dull tympany* over the whole area of the tumor, like that of large intestine, and distinctly lower pitched than over small intestines in the right flank. (U, Fig. 2.)

Vaginal examination. Outlet relaxed, uterus far back, pressed down into the sacral hollow. Through the anterior vaginal wall the tense, smooth, spherical wall of the cyst is distinctly felt. This is easily rocked from side to side, being movable to the extent of several centimetres.

Urinary examination. Before operation, March 25, 1890. Urine (by catheter) clear, light orange-yellow color; faint aromatic odor; acid reaction; albumin a trace, showing distinct thin ring with nitric acid; sediment, light white flocculi considerable in amount. Microscopically: A few hyaline casts, also a few epithelial cells and leucocytes.

Operation March 26.

April 3d. Urine (by catheter) clear, light-orange color; sediment mucus, moderate amount; albumin faint trace. Microscopically: A few hyaline casts, some faintly granular.

21*st.* Urine (by catheter) color lemon; reaction, faintly acid; specific gravity 1021; sediment moderate, light flocculi of whitish mucus; albumin, very faint trace. Microscopically: A few leucocytes, a few hyaline and faintly granular casts; granular débris, and some amorphous urates.

DESCRIPTION OF OPERATION.—Abdominal section by Dr. Kelly, March 26, 1890, at 9.30 A.M. Chloroform narcosis, duration forty-five minutes; pulse before operation 80, after operation 92; duration of operation to the beginning of the closure of the abdominal incision twenty-seven minutes, to completion thirty-five minutes.

First incision in the linea alba 10 cm. (3⅞ in.) in length, later extended up through umbilicus to 15 cm. (5⅞ in.); abdominal walls thin; at the second cut the peritoneum was laid open, exposing the mottled, reddish-white surface of a large sac, which was punctured and a large ovariotomy trocar thrust in, discharging six litres (about twelve pints) of thin yellow pus. On pulling down the cyst to evert it, it was found to be universally adherent. On attempting to strip off the adhesions around incision the hand was carried up under transverse colon, which was spread out over the anterior surface of the cyst.

The hand then carried into the cyst cavity revealed its enormous extent, reaching from right iliac fossa to floor of the pelvis high up into the left hypochondrium.

A calculus about 15 by 4 mm., dumbbell-shaped, was found in the tumor lying on the pelvic floor, suggesting at once renal origin.

The process of stripping was continued, gradually bringing the tumor forward, and dissecting it out of its bed, by using the fingers with a shearing movement through an opening made in the peritoneum to the inner side of the left descending colon. Strong adhesions everywhere bound the mass to the cellular tissue; these offered more difficulty on account of the enormous size and superficial extent of the tumor than from their density. One adhesion 2 cm. (¾ in.) in breadth, binding the tumor down to the psoas muscle in left flank, was separated, leaving bunches of muscular fibre on the tumor.

Up to this point the time was chiefly occupied in releasing this vast sac from its bed, extending like saddle-bags across the vertebral column.

The chief difficulties in the operation were now encountered in pulling the tumor down from above, high up under the left ribs and the diaphragm. On rolling the tumor over and exposing its posterior surface, a large vein 1 cm. (0.39 in.) in diameter was ligated. Parallel and to the inner side of this lay the ureter, which appeared sound and was not dilated; this was ligated at a point about 8 cm. (3+ in.) from the kidney.

After separating a number of adhesions from around the upper portion of the kidney, the whole mass was finally drawn down attached only by the hilum and its vessels; here a band 4 by 2 cm. (1½ by ¾ in.), including all renal vessels, was tied in five bundles and the kidney completely separated and removed.

At the time of emptying of the cyst, and again at its removal, the pulse became thready. Eight hypodermatics of brandy (each ♏xxx.) were given. Immediately upon removal of cyst 1.5 litres (3 pints) of hot water, 46° C. (115° F.), were poured into the abdominal cavity; its stimulant effect was at once apparent in the marked improvement in the heart's action, respiration, and general appearance. This was at once sponged out and more hot water poured in and sponged out from pelvis, right groin, over and under liver, around spleen, stomach, and small intestines, leaving the abdomen dry.

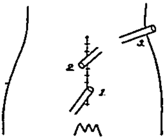
In the left iliac fossa the cellular tissues overlying the peritoneum had so fallen together as to practically separate the vast exposed surface from the peritoneal cavity.

On account of the shock and depression ether was substituted during the operation for chloroform, the latter being resumed at a later stage on account of the quieter narcosis.

The opposite kidney was examined and found to be somewhat enlarged (functional hypertrophy?), but apparently sound.

Drainage. With two fingers within acting as a guide, a scalpel was thrust through the skin, subcutaneous tissue, and muscles of the left flank posteriorly, half way between the crest of ilium and small ribs; through this hole a large glass drainage-tube was passed into the bed of the kidney, thus draining the cellular tissue beneath the peritoneum. (Fig. 4, No. 3.)

FIG. 4.



Shows the position and direction of the three drainage-tubes used. 1, Draining the pelvis; 2 and 3, draining the left flank.

The abdominal incision was closed with deep and superficial silk sutures, after insertion of two glass drainage-tubes in the median line, one at the lower angle of incision, extending into the pelvis, the other just below the umbilicus, extending over into the left flank. (Fig. 4, Nos. 1 and 2.) Dressings and abdominal bandage were applied, and the patient immediately put to bed and stimulated by hypodermatics of brandy and strychnine and external heat.

Convalescence. The patient was restless the afternoon and evening of the day of the operation, and had considerable pain the same night, requiring a single dose of morphia (one-quarter grain) hypodermatically.

The daily notes were as follows:

March 27. 12 M. Abdomen dressed; capillary gauze plugs in all three tubes saturated with dark, bloody discharge; there has been free discharge from drainage-tube in left flank; an area of cotton over tube size of hand stained by bloody discharge. There has been a very small amount of discharge from the other tubes in the line of the wound. Abdomen flat and flaccid; no nausea or vomiting; general condition excellent.

28th. 12 M. Abdomen dressed; all drainage-tubes removed; plugs in tubes stained with bloody discharge; very slight amount of bloody fluid swabbed from tube in left flank and lower one in abdominal cavity. Abdomen scaphoid; voided urine naturally last evening.

30th. Complains of "pain in bowels" this morning; has had five stools, large and small, during the past twenty-four hours.

31st. Doing well.

April 2. Color of skin of body losing its bronzed hue.

4th. Abdomen dressed; all sutures removed; union perfect, primary throughout, without any suppuration whatever; wound in flank made for drainage-tube healed also primarily. Abdomen flat, walls flaccid, wrinkled.

The patient had from this time on a rapid convalescence, uninterrupted by any bad symptoms.

She was nourished during the two days following operation mainly by nutritive enemata, with small quantities of coffee, and stimulated by hypodermatics of brandy, *p. r. n.*

On the second day small quantities of milk were given at frequent intervals, and on the fourth day she was given soft diet (toast, cornstarch, etc.).

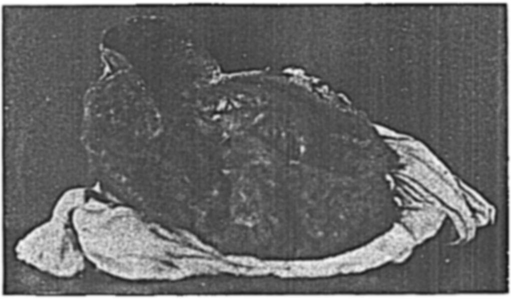
The temperature was 37.7° C. (100° F.) on the day after operation; from that time until the seventh day it ranged between 36.7° C. (98° F.) and 37.5° C. (99.5° F.); after which it remained normal, the pulse being correspondingly low. The patient sat up out of bed for the first time three weeks after operation, and was discharged from the hospital April 26, 1890, feeling perfectly well. She returned to her home in southern Delaware, whence she writes under date of February, 1891, that she 'feels splendidly; in fact, never felt better in her life."

PATHOLOGICAL EXAMINATION.—The tumor was examined in the pathological laboratory of the Johns Hopkins Hospital, and the following report made:

Abstract of report. It was only after repeated examinations that the tumor could be positively identified as a renal cyst. It was found to be made up of numerous sacs or cysts; the largest of these was twenty centimetres in diameter; this opened into a small sac with which most of the other sacs communicated. The collection of small cysts determined the congenital cystic nature of the kidney; the small sac with which they communicated was thought to have been originally the pelvis of the same, and the largest cyst the hydronephrosis consequent upon the obliteration of the ureter. The examination in detail was as follows:

The shrunken specimen presented for examination is a large sacculated mass, somewhat the shape of an enormous kidney. Its longest measurement is 44 cm.; width, 18 cm.; thickness, 7 cm.

FIG. 5.



Photograph of tumor lying upon a cloth, showing interior of large cyst.

The surface of the mass is covered with adhesions of fibrous and adipose tissue, but is in general smooth and glistening. It is composed of cysts of various sizes, several of which project from the surface. At a point on the concave surface which might correspond to the pelvis of the kidney, there is considerable adipose tissue, and a large artery which measures 2 cm. on the inner circumference enters here together with a large vein. Below the entrance of the artery and vein is another vessel corresponding to the ureter; 5 cm. of this remains attached to the tumor. This vessel is rather small; it measures 9 mm. in circumference and its lumen is patent. A mass of striated muscle of coarse fibre, measuring 2 cm. in thickness and 4 cm. in length, is attached to the upper and posterior part of the tumor. No communication between the ureter and the interior of the cysts can be made out.

At the time of the operation the largest cyst was opened and about a gallon of opaque, yellowish-white puriform fluid of creamy consistence escaped; also, a hard, angular mass, irregular in outline, 1½ cm. long and 1¾ cm. broad. The remaining cysts whose contents had not been evacuated by opening the large cyst were subsequently incised. These contained fluid of varying character. In one sac, about the size of a small orange, the fluid was clear; in another it had a reddish-brown color; and in the remainder it resembled the fluid of the large sac. The sacs composing the tumor vary in size; the longest, which composes about one-half of the tumor, measures 20 cm. in diameter, and communicates by a rounded opening, 1 cm. in diameter, with a small sac, with which most of the other sacs likewise communicate. The thickness of the wall of the largest cyst averages ⅔ mm.

Ridge-like projections of the wall protrude into the interior of the cyst for a distance of ⅓ cm. In the ridges can be seen a little grayish-red tissue, resembling renal tissue, and in the thicker parts of the wall, elsewhere, a little of the same kind of tissue can be seen; but, in general, the cyst wall seems to be composed of dense-gray, laminated fibrous tissues. The inner surface of the cyst is in part grayish, glistening, and smooth; but the greater portion of it is opaque, somewhat roughened, yellowish and reddish in color, and has small bloodvessels ramifying over it. The remaining cysts have in general a smooth, grayish, glistening inner surface, but in some places they likewise present a rough, opaque, yellowish deposit, somewhat resembling atheroma.

These remaining cysts likewise have projecting ridges from their walls, in which a little of the supposed renal tissue can be made out, but in general the walls are simply fibrous. They vary in thickness from $\frac{2}{5}$ mm. Most of them communicate by openings averaging 5 mm. in diameter with the large cyst or with the small sac above mentioned, into which the large cyst opens. Two of these cysts are completely closed, although they present cicatrix-like depressions, corresponding doubtless to the former openings, and in one instance a very fine probe can be passed through a minute opening in the cicatrix into an adjoining cyst. The largest cysts contain a few small, irregular, hard masses, $\frac{2}{5}$ mm. in diameter.

No communication between the supposed ureter and the cyst in the concave side, into which most of the cysts opened, could be found. Microscopical examination of fresh frozen sections of wall of largest cyst showed no definite kidney structure. Frozen sections of supposed ureter are confirmative. Scrapings from cyst wall, examined fresh, show leucocytes and compound granular bodies. No definite epithelial cells made out.

One litre of contents of tumor received into sterilized flask directly from trocar presents a yellowish-white, pus-like appearance, of creamy consistence. Sp. gr. 1.030. Reaction, very faintly alkaline. Markedly albuminous, coagulating on heating in test-tube. On standing twenty-four hours, the uppermost portion somewhat less dense in appearance, though there is no distinct sediment.

On microscopical examination it contains numerous compound granular bodies, multinuclear pus-cells, generally fatty degenerated cholesterin plates, and here and there highly refractive non-nucleated disks, resembling the "Drysdale" ovarian cell; also, fine acicular fatty crystals, singly and in groups.

Cover-slip preparations with aniline stains show no bacteria, though the granular matter appears in some instances to resemble micrococci. Four Esmarch's tubes (agar-agar) stab and smear inoculations made from cyst contents, placed in thermostat, developed no growths.

On further examination of hardened specimens, the yellowish material adjacent to the second largest cyst is almost certainly the greatly altered supra-renal capsule; it consists of epithelial cells which are large and finely granular, some of them containing large drops of fat. They are more or less arranged in masses, which are oblong, the longest diameter pointing to the centre of the mass, separated from one another by a connective tissue rich in cells. Among these epithelial cells there are large pale cells, with a horseshoe nucleus filled with granules, evidently compound granular cells. Adjoining and in the line of the section is a mass of dense connective tissue with a few cells containing numerous vessels. About these vessels is a certain amount of cell infiltration. Adjoining this is a mass similar to that first described, in which the bloodvessels are very abundant. This consists of epithelial cells, some of them of the ordinary shape and appearance of cells of the supra-renal capsule; others very large, pale, and granular. These cells lie in spaces smaller than those in the first-described mass; most of them very pale and granular. In the tissue there is a great deal of hemorrhage, which gives the microscopic appearance of pigmentation. Next comes a mass of dense connective tissue with few cells, containing glandular tissue and ducts lined with low columnar epithelium.

In the lumen of these a mass of more or less hyaline and granular material stained faintly with eosin. On the exterior of the section is a small portion of what is evidently the cyst wall. This is lined with a layer of vascular tufts, with nowhere complete lining of epithelium. The edge of it is composed of rather loose granulation tissue, with large, swollen cells. Here and there on the surface of this a few epithelial cells can be seen.

The second section shows about the same relations. On one end the cyst wall covered with papillæ; back of these a granulation tissue containing but few cells; and still back of this a dense connective tissue containing few cells and enclosing glandular tubules, which frequently contain colloid material; the tubules sometimes rather indefinite in shape and representing rather masses of epithelium surrounded by connective tissue. Past this, again, comes the loose connective tissue containing epithelial cells, which were considered to be the remains of the supra-renal capsules. There is a marked endarteritis everywhere, some of the arteries being entirely closed. The only evidence of kidney structure is found in the upper portion of the cyst wall, probably corresponding to the areas of kidney cortex. In the walls of the cysts themselves no definite layer of epithelium can be made out, though here and there, between the papillary projections, a few epithelial cells are seen. The connective tissue is general throughout the mass; it is dense and cicatrical, with but few cells. Here and there are areas of intense infiltration with round cells. In several places in the cyst are points which appear brownish to the naked eye; on microscopical examination of this part these are large round openings in the midst of a dense tissue, which contains few or no cells; these openings are lined with a highly refractive brown material, which forms a distinct layer around the openings, and evidently is of a crystalline character. The surface toward the lumen is broken and irregular; it is filled with lines radiating toward the centre, and is darker and more solid at the periphery. Within these masses, completely filling up the lumen, are masses of cicatrical tissue, which come from the surrounding tissue, penetrating between the crystalline masses. These structures are evidently to be regarded as arising from altered renal epithelium. They resemble most in color masses of uric acid, and have the same radiating lines which masses of these sometimes show.

The tumor might be regarded as simply a hydronephrosis, but another explanation, more probable than hydronephrosis, is to regard the whole as originally a congenital cystic kidney. In this case the small cyst into which the ureter and most of the larger cysts opened should be regarded as the pelvis of the kidney. The larger cysts of which the whole tumor is composed were either the original cysts of the kidney, or they have arisen from the continual dilatation of the cysts with rupture into one another. This would explain the various ridge-like projections on the wall. There might be a hydronephrosis in connection with this.

The supra-renal capsule has evidently been incorporated into the wall of the larger cyst in the upper portion of the kidney. Nearly all of the renal tissue was destroyed in the process, only here and there small remains of tubules can be found, which in most cases have undergone the peculiar transformation of their epithelium into the masses referred to.

FAT-DIGESTION.

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During the spring and summer of 1890, in the Berlin Physiological Laboratory, I made a study of the fat-splitting properties of pancreatic juice. I read a paper on this subject before the physiological section of the tenth International Medical Congress, and published a detailed account of my work in the *Journal of Physiology.* (*a*)[1]

It is my purpose in the present paper to show what application my published experiments (*a*) may have in explaining fat-digestion. That I may do this intelligently, it will be necessary to review our present knowledge of this subject.

C. Bernard believed that pancreatic juice had a twofold action on fats. In the first place, he said that when neutral oil and pancreatic juice were shaken together an instantaneous emulsion resulted, and in the second place, that the prolonged action of pancreatic juice on neutral oil would develop fatty acid. He did not in any way associate these two processes, and believed them to be due to entirely different properties of the juice; the emulsion being an instantaneous process and the fat-splitting occurring only after considerable time. And these two processes are described as separate and distinct properties of pancreatic juice in some of our most recent text-books. But some of the more recent German books have, upon the observations of Brücke and Gad, taught the relationship of these processes. From the fact that the addition of fatty acid to neutral oil makes the mixture readily emulsible in an alkaline liquid, they infer that the emulsion of fats in pancreatic juice, as observed by Bernard, is not due to an emulsion ferment, but is

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[1] Cambridge, Eng., April, 1891. The mark (*a*), which occurs frequently in the text, always refers to my published experiments in the Journal of Physiology.